

# Leukaemia Section

## Short Communication

## Lymphomatoid papulosis (LyP) with 6p25.3 rearrangement

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### Abstract

Review on lymphomatoid papulosis (LyP) with 6p25.3 rearrangement, with data on clinics, and the genes implicated.

### Clinics and pathology

#### **Disease**

Lymphomatoid papulosis (LyP)

#### **Phenotype/cell stem origin**

Mature (peripheral) T cell.

#### **Etiology**

No etiologic factors are known.

#### **Epidemiology**

All reported cases occurred in elderly adults (67-88 years, mean 75 years) with strong male predominance (9M:2F) (Karai et al., 2013).

#### **Clinics**

The lesions are usually restricted to a single body area.

Affected sites included the head and neck, upper torso, and extremities.

Lesions ranged in size from 0.3 cm to 1.0 cm in diameter and had variable scale but no ulceration.

No patient had disseminated cutaneous disease or clinical evidence of extracutaneous disease.

### **Pathology**

The histological features are remarkably consistent and different from the other types of cutaneous LyPs.

Salient features include: prominent dermal nodule with the overlying epidermis showing pagetoid reticulosis-like histological changes.

Marked periadnexal involvement is sometimes present. Tumor cells are mostly small to medium-sized with markedly irregular nuclei.

Lesions show high mitotic rate and the presence of frequent apoptotic bodies.

Necrosis is absent.

No significant amount of eosinophils and neutrophils are present.

### **Treatment**

No established treatment.

Most of the lesions involute spontaneously and without therapy.

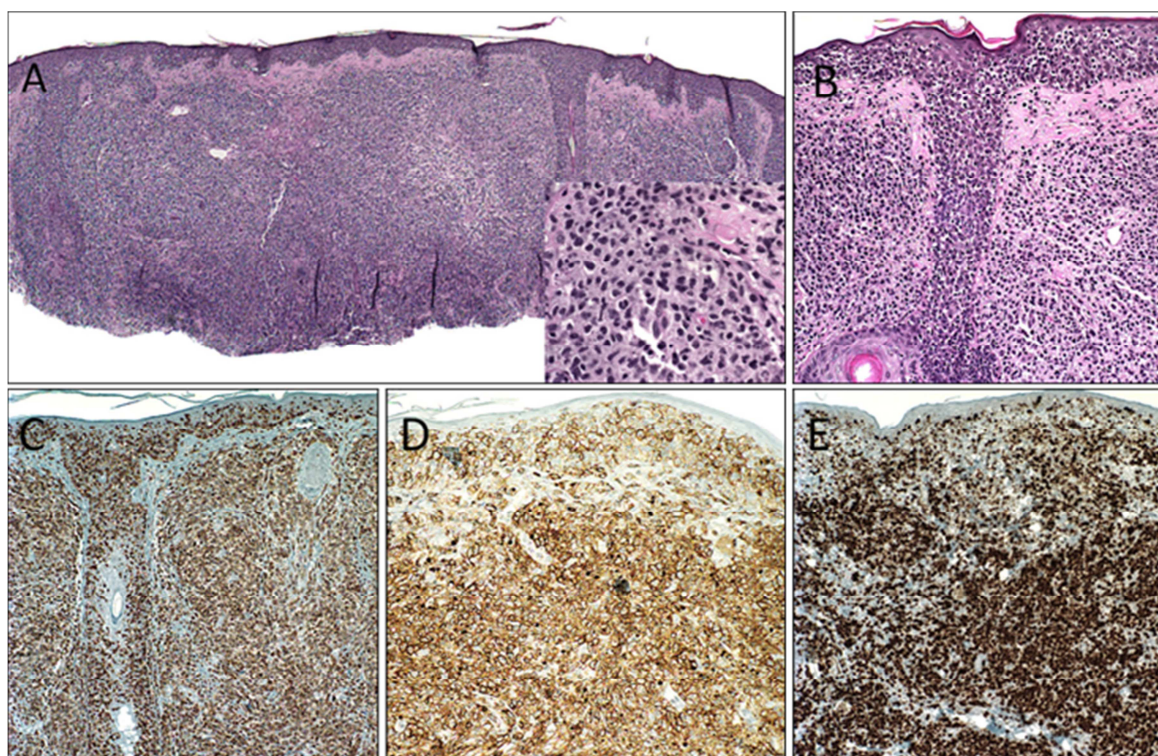
Consideration to radiation, injection with kenalog or excision can be made depending on individual presentation.

### **Evolution**

Similar to other types of LyP, recurring-remitting course.

### **Prognosis**

According to the series of 11 patients the prognosis is good (Karai et al., 2013).



Lymphomatoid papulosis with 6p25.3 rearrangement. Low power H&E image shows a prominent lymphoid nodule in the dermis (A) with characteristic pagetoid reticulis-like epidermal involvement (B) at medium power. Inset in (A) shows high power image of atypical cells with small to medium nuclei and with the presence of frequent apoptotic bodies and mitotic figures. Immunohistochemical stains reveal a CD3 and CD30 positive phenotype with high proliferative ratio depicted by Ki-67 (C, D, and E, respectively).

## Cytogenetics

### *Cytogenetics morphological*

Karyotypic findings have not been reported.

### **Probes**

See Karai et al., 2013.

### **Additional anomalies**

Unknown.

### **Variants**

Unknown.

## Genes involved and proteins

### **Note**

The breakapart probe used to identify 6p25.3 rearrangements in LyP spans both the DUSP22 and the IRF4 genes.

The specific breakpoints have not been reported in LyP. In ALK-negative anaplastic large cell lymphomas (ALCLs), 6p25.3 breakpoints in or near either gene have been reported (Feldman et al., 2011). The partner locus was confirmed to be

7q32.3 in a single case of LyP tested, analogous to the t(6;7)(p25.3;q32.3) reported in ALCLs, but was not investigated in the remaining reported cases.

### **DUSP22**

#### **Location**

6p25.3

#### **Protein**

DUSP22 encodes a dual-specificity phosphatase that inhibits T-cell antigen-receptor signaling in T cells by inactivating the MAP kinase, ERK2.

The expression and function of DUSP22 in cases of LyP with 6p25.3 rearrangements have not been reported.

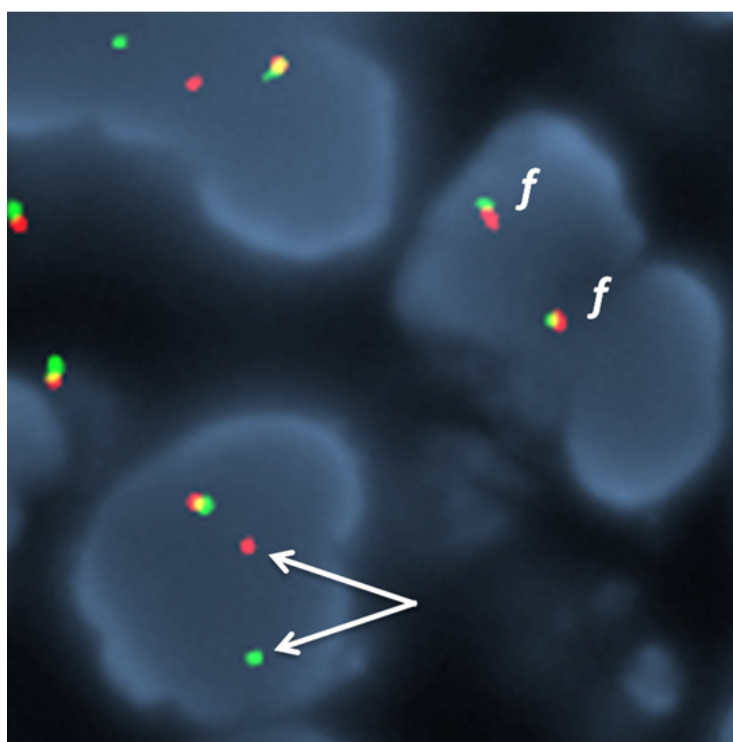
### **IRF4**

#### **Location**

6p25.3

#### **Protein**

IRF4 encodes a transcription factor, IRF4/MUM1, critical in lymphocyte activation and differentiation. Although IRF4/MUM1 is expressed in most cases of LyP, the expression and function of IRF4/MUM1 in cases of LyP with 6p25.3 rearrangements have not been reported.



Breakapart FISH for the DUSP22-IRF4 locus on 6p25.3 demonstrates abnormal separation of the red and green signals in the cell in the lower left (arrows). A cell with a normal signal pattern (two fusion signals, f) is seen in the upper right

## Result of the chromosomal anomaly

### **Hybrid gene**

#### **Note**

Not known.

### **Fusion protein**

#### **Note**

Not known.

## References

Feldman AL, Dogan A, Smith DI, Law ME et al.. Discovery of recurrent t(6;7)(p25.3;q32.3) translocations in ALK-negative anaplastic large cell lymphomas by massively parallel genomic sequencing. *Blood*. 2011 Jan 20;117(3):915-9

Karai LJ, Kadin ME et al.. Chromosomal rearrangements of 6p25.3 define a new subtype of lymphomatoid papulosis. *Am J Surg Pathol*. 2013 Aug;37(8):1173-81

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